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ABSTRACT

The paper reports on studies involving children having the rare chronic skin diseases of hemangiomas and epidermolysis bullosa (characterized by easy blistering). One study compared the self-concept and psychosocial development of young (mean age 46 months) children (N=19) with hemangiomas with 19 children without hemangiomas. Findings indicated few differences in self-concept or psychosocial development between children with and without hemangioma lesions. A national survey of children with epidermolysis bullosa found that 15.8% were receiving special education assistance under Public Law 94-142, the Education for All Handicapped Children Act with 82% receiving services in regular classes. Environmental accommodations and adaptive devices were used. Although 48% reported no social problems, 32% felt there was some peer rejection as a result of their condition. An investigation into developmental characteristics of five young children (ages 3-6) having mild to severe epidermolysis bullosa found a wide variance in overall developmental status with motor skills delayed in proportion to the severity of the condition. Includes 18 references. (DB)

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CHILDREN WITH RARE CHRONIC SKIN DISEASES:

HEMANGIOMAS AND EPIDERMOLYSIS BULLOSA

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Abstract

The purpose of this paper is to present information on two independent studies that were conducted involving children having rare chronic skin diseases: hemangiomas and epidermolysis bullosa. Depending on the severity of these skin disorders, they can have far reaching implications for normal cognitive, motor, language, and social-emotional development. Not only can these conditions be devastating for the child, but caring for the child can create psychological, social, and economical stress for the parents. This presentation, which will be divided into two parts, will provide information about the characteristics of each skin disease, implications for normal development, and implications for the classroom.

INTRODUCTION

Hemangiomas and Epidermolysis Bullosa are two rare chronic skin diseases that have implications for normal development, family functioning and educational programming. Because of the small number of chronically ill children and the rarity of many diseases, the needs of these children as individuals and as a group have been neglected by our society. The purpose of this session is to present information on two independent studies that were conducted involving children having rare chronic skin diseases: hemangiomas and epidermolysis bullosa. Depending on the severity of these skin disorders, they can have far reaching implications for normal cognitive, motor, language, and social-emotional development in young children. Not only can these conditions be devastating for the child, but caring for the child can create psychological, social, and economical stress for the parents. This presentation and paper, which will be divided into two parts, will provide information about the characteristics of each skin disease, implications for normal development, and implications for the classroom.

Hemangiomas

The relationship between a physical anomaly and a child's development of self-concept and personal relationships has been a concern to many researchers (DeGrella & Green, 1984; Lefebvre & Munro, 1978, 1986; Moncada, 1987; Sorell & Nowak, 1981). However, the understanding of the psychosocial development and self-concept of children with hemangiomas, a common but unique cutaneous vascular defect, remains limited.

Hemangiomas erupt during the first few weeks of life and grow quickly for 6-12 months. Although lesions usually begin to involute during the second year, and 90% of the hemangiomas regress by the age of seven (Cohen, 1987; Hurwitz, 1981; Lister, 1938), no study has assessed the psychosocial health, self-concept development, or family functioning of children with self-limited but transiently disfiguring hemangiomas.

The purpose of this study was to assess the self-concept and psychosocial development of young children diagnosed with hemangiomas as compared to young children without lesions. In addition, to compare the

family functioning of children who have hemangiomas and those who do not.

Method

Subjects

Subjects included 19 patients with hemangiomas, 15 girls and 4 boys whose ages ranged from 35 months to 70 months ($x = 46.63$) and their mothers. Patients with hemangiomas were identified through the Pediatric Dermatology Clinic at the Children's Hospital of Pittsburgh. All patients were referred for dermatologic evaluation by a private pediatrician or dermatologist. A control sample consisting of 15 girls and 4 boys without tumors ranged in age from 36 months to 64 months ($x = 45.89$). Subjects in the control group were recruited from day care centers, preschool parent groups, and private pediatric practices. Hemangiomas involved the face, head or neck. All patients were assigned a degree of lesion involvement based on a five-item, 3-point rating scale (see Figure 1). Tumors were rated by degree of color, elevation, location, diameter, and surface area. Scores for each individual were added across the five categories for a

maximum lesion involvement index of 15. Involvement index scores ranged from 5 to 15 ($x = 9.9$). On the average, hemangioma lesions were diagnosed at 1.5 months. All subjects were Caucasian. Prior to data collection the study was approved by the Human Rights Committee at the Children's Hospital of Pittsburgh and the Human Subjects Review Board at Kent State University. Informed consent was obtained from participating parents.

Instruments

Data was collected from parents and children in both groups during a home interview. Parents completed the Achenbach Child Behavior Checklist (CBCL) as a measure of the child's psychosocial functioning (Achenbach, T. M., & Edelbrock, C. S., 1983). The CBCL includes a listing of behavioral problems (e.g., child demands a lot of attention, likes to be alone, feels too guilty, gets in many fights, etc.) that are rated by the parent on a 3-point response scale. Parents also completed the Family Functioning Index (FFI), a self-report measure of marital satisfaction and communication.

Each child was given the Joseph's Pre-school and Primary Self-Concept Test (JPPST), (Joseph, J., 1979). The JPPST is a measure designed to identify a child's self-perception, behaviors, views of others, and feelings of worth and satisfaction attached to these perceptions. The JPPST consists of three dimensions: (1) Significance, (2) Competence, and (3) General Evaluative Contentment.

Results

Analyses to determine differences between patients with hemangiomas and those without lesions on the CBCL and JPPST were conducted using Hotelling's T^2, a multivariate analysis of two groups of subjects on several dependent variables. Pearson product-moment correlations were computed only for the groups of patients with hemangiomas relating index of lesion involvement with psychosocial functioning, self-concept, and family functioning. Pearson product-moment correlations were also used to relate self-concept and psychosocial development with family functioning.

Self-Concept

Results of the Hotelling's T^2 indicate a difference between the two groups on the self-concept measure (JPPST), $F(3,33) = 3.71$, $p<.05$. Follow-up univariate analyses indicate a difference between the groups for the Significance dimensions (Table 1). Children with hemangiomas more often identified with pictures of a child playing alone, less often picked themselves as the child mom and dad liked, and more often associated with the child who had no toys. No difference was found between the two groups for the Competence of Evaluative Contentment dimensions.

Psychosocial Functioning

Results of the Hotelling's T^2 conducted on the CBCL indicated no difference with respect to the psychosocial functioning of children with hemangiomas and children without lesions, $F(3,33) = .73$, $p>.05$. Children in both groups were rated on the CBCL within the range of acceptable psychosocial development. However, the patient with the most lesion involvement was rated as having the most psychosocial problems and lowest self-concept across both groups.

Table 1

Means of All Dimensions on the Self-concept Measure for Children in the Target and Control Group

Item	Target (n=18)	Control (n=19)	t tests
Significance Dimension			
Play with friends/alone	1.?6	1.94	-2.57*
Teacher likes/not like	1.?3	2.00	-1.76
Parents like me/sibling	1.57	1.94	-2.00*
Get spanked/candy	1.77	1.89	- .59
Lots of toys/not toys	1.57	2.00	-2.39*
Lots of candy/little candy	1.61	1.84	-1.01
General Evaluative Contentment (GEC) Dimension			
Clean/dirty	1.89	2.00	
Like name/Not like name	1.57	1.57	
Smiles/cries	1.78	1.94	
Likes house/Not like house	1.73	1.89	
Competence Dimension			
Says a lot/Not a lot	1.36	1.47	
Runs slow/Runs fast	1.57	2.00	
Jump high/Not jump high	1.57	1.63	
Win game/Lose game	1.63	1.94	

Note. Univariate tests were not calculated for GEC and Competence dimensions since no differences were found on the multivariate test.

* $p<.05$.

Family Functioning

Pearson product-moment correlations were used to relate self-concept and psychosocial functioning with family functioning for the target and control group followed by *z*-tests to determine the difference between the independent correlations. No significant difference was found between the target and control group regarding the correlation of self-concept and family functioning (see Table 2). The *z*-test for testing the difference between independent correlations for psychosocial development and family functioning did approach significance, $z = 1.95$, $p<.05$ (see Table 2).

Lesion Involvement

According to the Pearson product-moment correlation, no significance was found relating index of lesion involvement with psychosocial functioning or self-concept (Table 3).

Discussion

Evaluation of the children's self-concept and psychosocial development indicated few differences between young children with hemangiomas and those without lesions. Children with hemangiomas perceived

Table 2

Differences Between Correlations of Self-Concept and Psychosocial Functioning with Family Functioning

Target Variable	Control (n=15)	(n=18)	z value
Self-Concept	.21	.64**	-1.40
Family Functioning	-.11	-.70**	1.95b

Note. None of the differences between correlations achieved statistical significance.

*p<.05.

**p<.01.

Table 3

Correlations Between Index of Lesion Involvement, Self-Concept, Psychosocial Functioning, and Family Functioning

Variable	Involvement Index	p
Self-Concept	-.37	.12
Psychosocial Functioning	.35	.13
Family Functioning	.06	.82

Note. None of the correlations reached statistical significance.

themselves less positively on the Significance dimension of the self-concept test, which is a measure of children's perceptions of how others value them. A possible explanation for the negative response to this dimension is that the young children with lesions can not hide the tumors from significant persons.

Mothers of children with hemangiomas reported no greater number of problem behaviors than did mothers of children in the control group. Mothers in both groups rated the children equally competent with respect to psychosocial behaviors as measured on the CBCL. This result is encouraging for parents just beginning to deal with the development of tumors. It appears that children with hemangiomas do not behave differently from same aged children who do not have lesions. This is encouraging since children with physical conditions often demonstrate more behavioral difficulties. Drotar, Doershuk, Stern, Boat, Boyer, and Matthews (1981) explain that the anxieties children experience with their bodies and altered physical appearance is often reflected in an increase of psychosocial problems. Children in this study who have hemangiomas

apparently are not experiencing anxieties related to the condition and are appropriately managing behavioral responses to a visible condition.

Furthermore, no difference between the two groups on psychosocial functioning relates to Dreikurs' (1978) application of Adler's (1970) organ inferiority. It may be that children with hemangiomas are making the most of having a biologically endowed condition. It is not what one has in heredity endowment but what one does with it that is important. In this study children with hemangiomas appear to be managing behavioral responses to their hemangiomas.

Parental comments were directed toward the child's association with other children, particularly at the onset of school. Mothers identified that comments from other young children so far were inquisitive (e.g., how did they get that, does it hurt). However, mothers speculated that these inquiries would not remain as positive once the child began school, a concern that might warrant removal of the tumor, if medically feasible, before kindergarten or shortly thereafter even if there is the risk for scarring. Studies of

other facial deformities including cleft palate (Abroms, 1985) suggest that the earlier corrective surgery can be performed, the better the chances of the development of a healthy self-concept. Of course, surgery is not a panacea as indicated in surgical intervention studies conducted by Lefebvre and Munro (1978, 1986). Parents expect surgery to resolve any tensions related to having a child with a facial difference. Realistic expectations need to be set with respect to postoperative outcomes. Unrealistic expectations can disappoint parents. Such response can further negate the child's emerging self-concept.

This study was limited to one age group. The child's development following entrance into formal education and establishment of extended peer relationships was not examined. Additionally, the perceptions of siblings related to living with a child who has hemangiomas was not addressed.

Other than the child, parents were the only source of data investigated in this study. However, many others may affect the child's development, including

physicians, relatives, neighbors, peers, social workers and teachers.

Although few differences were found between children with and without hemangiomas, physicians and other health care professionals need to continue (1) disseminating appropriate medical information regarding lesion development and resolution, (2) monitor the child's social/behavioral progress and lesion development with respect to possible surgical intervention, (3) provide parental support with photographic progress, and (4) recruit "experienced parents" (parents of children with resolved or nearly resolved lesions) to provide testimony that, even though it does get worse before it gets better, lesions can naturally resolve.

In the future we would like to expand this pilot investigation. This can be accomplished by securing a larger sample and including older children. Although hemangiomas usually resolve without treatment, a good result may not occur until later childhood or adolescence. Further data will help the practitioner to determine the optimal point in the child's

development to consider surgical intervention if necessary.

Summary

In this study the psychosocial development and self-concept of young children with hemangiomas was compared to young children without tumors. This study was limited to preschool aged children who have visible hemangiomas. It was found that children with hemangiomas are not demonstrating problems related to psychosocial development but do experience some difficulty with how others value them. This suggests the need for continued assessment and support for child and parent, particularly during the months of rapid lesion development. An understanding of how children respond to spontaneous resolution or medical intervention are concerns for future study.

EPIDERMOLYSIS BULLOSA

Epidermolysis bullosa (EB) is a rare, chronic condition which can occur in many forms. It is characterized by the formation of blisters which usually result from the slightest degree of pressure or trauma to the skin. Although the prevalence of

epidermolysis bullosa is not precisely known, it has been estimated that the condition may occur as frequently as once in every 50,000 births for the dominant types and once in every 300,000 births for the recessive types (Artnak, et al., 1981). It is believed by the Dystrophic Epidermolysis Bullosa Research Association, Incorporated (National Institute of Health, 1984) that approximately 50,000 persons in the United States have the condition.

At the present time, medical research has not been able to discover the exact cause of the condition nor link it to genetic markers, such as ABO blood types, RH type, or sex (Artnak, et al., 1981). Histopathologically all forms have a cleft between the epidermis and dermis and can be classified as nonscarring (epidermolysis bullosa simplex-generalized, epidermolysis bullosa simplex-localized, and junctional epidermolysis bullosa) and scarring types (dominant dystrophic epidermolysis bullosa and recessive dystrophic epidermolysis bullosa).

Koebner and Dowling Meara are two types of epidermolysis bullosa simplex-generalized. Of these

two types of epidermolysis bullosa simplex-generalized, the Dowling Meara variant is more serious and life-threatening during the neonatal state. The Koebner variant is usually present at birth or becomes apparent during infancy as the child begins to crawl and walk. With the generalized types of epidermolysis bullosa, blisters can occur over the entire body's surface if pressure or injury occurs to the skin. As a result of mechanical friction, caused by manual labor and physical activities, blisters are most likely to form on fingers, toes, palms, soles, and the back of the heel and neck. These blisters heal without scarring. Involvement of the mucous membranes, fingernails and toenails may also occur. Blistering is more severe if the skin gets warm and wet from perspiration. Epidermolysis bullosa simplex-generalized persists throughout life, but appears to diminish in severity after puberty (Gedde-Dahl, 1971).

Epidermolysis bullosa simplex-localized, often called Weber-Cockayne, is characterized as a condition which has the blisters forming almost exclusively on the hands and feet. This type usually develops in

childhood as the child begins to use the hands to manipulate objects and becomes more mobile. The blisters heal without scarring.

Junctional epidermolysis bullosa can appear in two forms: mild and severe. Blisters, which seem to occur spontaneously, are present at the time of birth. Large areas of the body's surface, particularly the trunk and legs, become denuded of skin. The skin heals without scarring, but the skin may be tight. The condition may diminish after the first year of life if the infant survives. Infants may die as a result "of profound loss of fluid, overwhelming infection or complications resulting from blistering in the gastrointestinal, respiratory or genitourinary tract" (National Institute of Health, 1984).

One of the scarring forms of the condition is dominant dystrophic epidermolysis bullosa, which is usually present at birth or early infancy. Mechanical trauma causes blistering to occur over the entire body or only to specific locations of the body, such as on the hands, feet, elbows, or knees. Severe scarring can occur, but does not seem to cause immobility or

deformity of the hands and feet. However, nails are often thickened, malformed, or destroyed (National Institute of Health, 1984).

Recessive dystrophic epidermolysis bullosa is the other scarring form of the condition. Mild to severe forms can occur. Present at birth, recessive dystrophic epidermolysis bullosa is characterized as having nearly all skin surfaces and mucous membranes (from mouth to anus) covered by blisters with large body surfaces denuded of skin. The esophagus can become scarred. This scarring can interfere with the passage of food, thus causing chronic malnutrition to occur. Scarring and deformity are very serious, with fingers and toes fusing together and becoming immobile. Scarring may also cause hands and arms to become fixed in a flexed position. In addition, the nails, teeth, and eyes are also involved (National Institute of Health, 1984).

Within each form of epidermolysis bullosa, the condition may be exhibited in various degrees of severity. Thus, a child with severe epidermolysis bullosa-simplex (nonscarring) may have more

complications than a child with a mild form of dominant dystrophic epidermolysis bullosa (scarring). In addition, each form of the condition has different implications for the individual's social, physical, and psychological development and adjustment. The manner in which the condition influences development and daily adjustment depends on its severity, the prognosis, the degree of limitation imposed, the necessity for physical care, and the visibility of the resulting deformity and scarring.

EB & Educational Research Findings

Two studies have been conducted that investigated the educational implications of epidermolysis bullosa for children and families. One national survey research study, conducted by Jones and Magnusson (1985), focused on educational provisions for children with epidermolysis bullosa and their families. Surveys were mailed to 225 families affiliated with the Dystrophic Epidermolysis Bullosa Research Association of America, Inc., with a response return rate of 72%. The ages represented a range from 3 to 29 and included grade levels from preschool through college.

Approximately 80% were between 5 and 19 years of age. The three major groups of epidermolysis bullosa were represented as follows: 49% - EB simplex, 3% - junctional EB, 25% - dystrophic EB, and 23% were not specifically named. Eighty-one percent of the children were described as having mild to moderate severity, meaning healing with or without scarring. Nineteen percent were described as have severe EB with poor healing with scarring, webbing, and other complications.

The survey findings revealed that only 15.8% were receiving assistance under the provision of Public Law 94-142. Children with epidermolysis bullosa were labeled by school districts as follows: 33% - health impaired, 27% - noncategorical, 20% - gifted, 19% - orthopedically impaired, 9% - learning disabled, 6% - speech impaired, 4% behavior disordered, and 1% - language impaired. Eighty-two percent of the school-aged children were in the regular classroom and 12% were in a special classroom or program (6% - resource room, 2% - self-contained classroom, and 4% - homebound instruction).

The survey revealed that certain environmental accommodations and adaptive devices were being used by the children. Sixteen percent of the school-aged children left the classroom prior to the end of the period to avoid being bumped in the hall; 13% of the children were in air-conditioning, and 15% of the children had the use of a teacher aide if necessary. Adaptive devices included typewriters, tape recorders, computers, pencil holders, use of cushions in class, duplicate sets of books for home use and extended time for tests.

In the area of socialization, 48% reported no problems. However, 32% felt there was peer rejection, and another 32% expressed difficulty in making friends.

A second study, conducted by Jones (1987), investigated the developmental characteristics of five young children (ages 3-6) having mild to severe epidermolysis bullosa. The developmental characteristics of the five children were determined through the use of the Denver Developmental Screening Test, the Diagnostic Indicators for the Assessment of Learning-Revised, and the Vineland Adaptive Behavior

Scales. The five children exhibited a wide variance in overall developmental status and developmental characteristics in the areas of language, motor, and social-emotional development. Language development tended to follow norm expectations; however, items requiring the identification and understanding of preposition were performed below expected age norms. Motor development tended to vary from norm expectations depending on the severity of the condition. As the severity level of the condition increased so did the number of motor skills performed below expected norm levels, particularly fine motor skills requiring manipulation of objects. Social-emotional development had the greatest variance from norm expectations, with a clustering of socialization items performed below expected norm levels. It was concluded by Jones (1987) that the condition resulted in a protective environment and/or lack of opportunity to play freely and make friends. Children who have severe forms of epidermolysis bullosa are often kept in safe, secure environments to protect them from injury and infection. For some children, the lack of physical strength,

endurance and gross motor skills may not make it possible for them to engage in physical childhood activities such as tag or riding bikes.

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